Acral lentiginous melanoma: Case studies from Kauai

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Cutaneous malignant is generally regarded as a skin cancer of Caucasians. However, one subtype, acral lentiginous melanoma (ALM), has an equal distribution among all ethnic groups and is the variant most common in non-Caucasians. In its early stage it is difficult to recognize and, unless diagnosed early, has a very poor prognosis. Herein, we present 3 cases of ALM seen on Kauai in 1990.

Introduction

Cutaneous malignant melanoma is increasing in incidence at an alarming rate. If this trend continues, it is estimated that the incidence of melanoma in all Americans born in the year 2000 will be 1:90[1]. This estimate apparently refers to Americans of all ethnic backgrounds, thus the incidence in Caucasian Americans will be substantially higher since melanoma is at least twice as prevalent in Caucasians as in non-Caucasians.

Melanoma is subdivided into 4 clinicopathologic types: Superficial spreading, nodular, acral lentiginous (ALM) and lentigo maligna melanoma.

ALM affects the glabrous or non-hair-bearing surfaces of the skin and includes the volar aspects of the hands and feet and the subungual regions. Superficial spreading and nodular melanoma may also affect these locations; however, ALM is the most common variant at these sites. Due to its somewhat concealed location, it may not be recognized until in an advanced stage.

ALM affects Caucasians and non-Caucasians to the same extent. However, the overall number of cases of melanoma is much less in non-Caucasians so the relative percentage of ALM is higher in this group.

We report on 3 cases of ALM all seen in patients of predominantly Filipino ancestry on Kauai during 1990.

Case Histories

Case 1 is a 66-year-old Filipina who presented to her family physician with what appeared to be an area of infection measuring 8 mm in diameter on the instep of the sole of her right foot. She was treated with antibiotics; however, the skin lesion did not resolve. The clinical impression was then changed to a traumatized plantar wart. Wisely, a skin biopsy was performed; a histologic diagnosis of ALM was made and the patient underwent a wide local excision with grafting (Fig 1). There was no evidence of metastasis and the patient is doing well.

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Case 2 is a 49-year-old Filipino/Hawaiian man who presented to his doctor with a 2 cm dirty ulcer on the heel of his right foot. The patient, unable to give a good history, stated that the ulcer had been present "for 10 years" and it bothered him when he drove his truck. A biopsy was interpreted as ALM (Fig 2). The patient is currently undergoing further treatment elsewhere.

Case 3 is a 32-year-old Filipina who presented to the emergency room complaining of tightness in her right foot while jogging. She stated that a "mole" had been present on her left 4th toe for the past year. She had lost 30 pounds in weight in the past 4 months and suffered from anorexia, nausea and vomiting during the past month. Physical examination revealed an ulcerated, dark-brown nodule on the dorsomedial aspect of her left 4th toe (Fig 3). She had multiple subcutaneous nodules measuring up to 4 cm in diameter over her body. A biopsy confirmed the clinical suspicion of ALM. CT-scan disclosed metastases to the eye and cerebrum. The patient died one month later. No autopsy was performed.

Discussion

The number of cases of melanoma increases every year. Approximately 27,600 new cases of melanoma are expected to be diagnosed in the U.S. this year[2]. The 5-year survival rate in the U.S. has improved from 43% in the 1930s to 83% in 1983[2]. Despite this improvement, mortality has more than doubled over the same time period due to the increase in the incidence. A 3- to 4-fold increase in incidence has been reported in the 20-year period 1960 to 1980; for example, in both Japan and Hawaii[3,4]. This increase pertains to all 4 subtypes of melanoma.

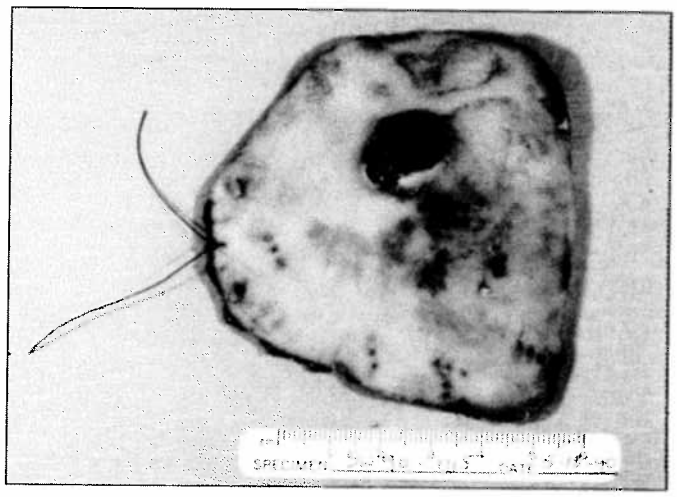

Fig. 1 - Case 1. Wide local excision of plantar ALM.

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In the mid-1970s, RJ Reed categorized ALM as a separate subtype of melanoma based on its anatomic sites of predilection and its distinct histology[5]. There is a great ethnic/racial variation in the percentage of melanomas that are of the ALM type. It is highest in Africans and African-Americans (upward of 76%) and lowest in whites (less than 10%)[6,7,8,9,10]. The remainder of the ethnic groups fall between these extremes. Of note is the fact that Chinese, Japanese, Southeast Asians (including Filipinos) and Polynesians are predominantly in the upper portion of this intermediate zone of distribution; with cases of ALM ranging between 29% to 74% of all cases of melanoma in these ethnic groups[4,6,7,9,10].

ALM has a peak age incidence in the seventh decade of life in man and in the sixth decade in women[1]. The other 3 subtypes of melanoma are linked to intermittent sun exposure and burning; however, the etiology of ALM is uncertain[11]. Neither trauma nor pigmented nevi appear to predispose to ALM[7,8,9,12].

Caucasians are most prone to develop melanoma and this predisposition is indirectly proportional to the amount of pigment in the skin. For instance, the incidence is highest in red-headed Caucasians who freckle easily. Blacks and other darker-skinned races have a higher percentage of distribution of melanomas on the palmar-plantar surfaces, although the actual incidence at that site is very similar for all races since skin in these areas is relatively depigmented. Thus, non-pigmented skin in whites and non-whites is the most likely to develop melanoma.

Plantar melanoma outnumbers palmar melanoma by a factor of 17:11[3]. In its early stages of radial growth, it appears as a variably pigmented, brown macule with subtly asymmetric, irregular, notched borders (Fig 4). This stage may last for several months or years; then, there occurs a rapid change to the vertical phase of growth, heralded by a dark nodule (Figs 1,3,5). Since glabrous skin can extend medially and laterally around the digits, ALM also may arise on dorsolateral surfaces of the hands and feet (Fig 3,5). Longitudinal dark-striations in the nail bed or perionychium (Hutchinson's sign) may herald a subungal ALM.

It is difficult to recognize ALM during the radial growth phase but excisional treatment at this stage offers an excellent prognosis. During radial growth, and in the later vertical growth phase, plantar ALM can be misdiagnosed as a nevus, plantar wart, corn, callus, infected site of trauma (with hemosiderin deposition), hematoma, pyofenic franuloma, or even a focal area of gangrene. A high index of suspicion is needed to make the diagnosis. Any atypical lesion of the plantar surface, especially one with irregular borders or ulceration, should be biopsied.

In the majority of cases, ALM has a distinct histologic appearance (Fig 2). There is marked hyperplasia of atypical melanocytes with long dendritic processes along the base of the epidermis. Unlike other subtypes of melanoma, there is usually little if any migration by the atypical melanocytes into the upper epidermis. There is thickening (acanthosis) of the epidermis and irregular, wide elongation of the rete ridges. In its vertical growth phase, a predominance of atypical spindle cells with increased mitoses extend downward into a desmoplastic dermis.

The prognosis in ALM compares favorably with other subtypes of melanoma if the depth of invasion is minimal[1,2,13]. However, ALM is usually diagnosed rather late due to its obscure location and ill-defined appearance; thus, it carries an overall poor prognosis. Blacks have a worse prognosis than whites with 5- and 10-year survival rates being 18% and 18% for blacks and 72% and 16% for whites[8,13]. The 5-year survival rate of ALM in Japan, according to one study, was 29% as compared to 39% for all melanomas[3].

Prognostic indicators in ALM are similar to that in other melanomas and include: Depth of invasion (Clark's dermal or subcutaneous level of invasion and Breslow's measured depth of thickness); ulceration (if present, it carries a worse prognosis); stage (involvement of lymph nodes, presence of distant metastates); and mitotic rate or index (an increased number of mitoses lowers the chance of survival)[2,14,15].

The depth of invasion is considered to be the prognostic factor of greatest significance. Both Clark level I (tumor confined to the epidermis and its appendages) and Breslow depth of invasion less than 0.76 mm carry a 10-year survival rate of greater than 95%[2]. According to the staging system of melanoma by the American Joint Committee on Cancer (that takes into account depth of invasion, lymph node involvement and metastases), the 5- and 10-year survival rates for Stage I (Clark level III or less; depth of invasion less than 1.5 mm; no lymph node or distant metastasis)

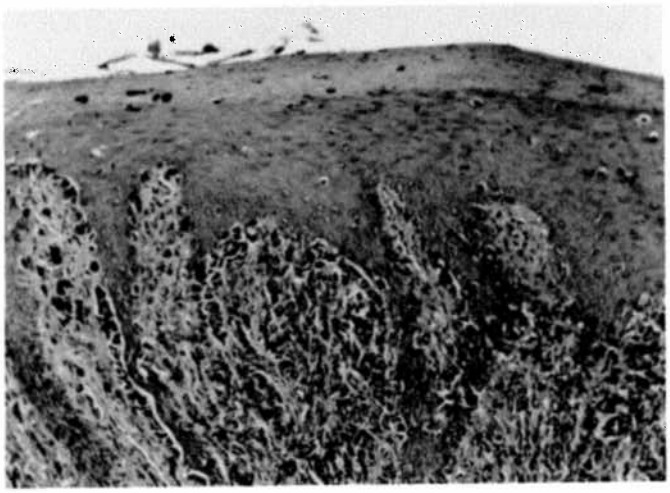

Fig. 2 - Case 2. Plantar ALM. High power. H&E stain.

Fig. 3 - Case 3. ALM of foot, dorsomedial extension.

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are 97% and 95% respectively[2].

The major drawback in using these prognostic criteria for ALM is that in most cases the level of invasion into the soles or palms is difficult to assess, since the normal increased thickness of the epidermis in these locations exaggerates the true Breslow depth of invasion. There also is poor demarcation between the papillary and reticular dermis at these loci with little, if any, subcutaneous fat present and thus assignment to the Clark level is usually arbitrary.

The treatment of ALM is wide, local excision or amputation[13,16]. Elective regional lymph node dissection (without clinical evidence of tumor involvement) is generally not indicated. Lymph node dissection is not recommended in the case of widespread metastases.

In advanced stages chemotherapy, immunotherapy (most noteworthy, interleukin-2), hormonal therapy (melanocyte-stimulating hormone targeting of cytotoxic agents), and fast neutron therapy are used in a few specialized treatment centers; however, the prognosis at these stages is generally very poor. Melanoma is generally regarded as radioresistant.

Early diagnosis and surgery offer the best chance of cure. Unfortunately, as illustrated in our case number 2 and 3, patients may not be aware there is a lesion on the sole of their foot and, if so, most often associate it with trauma. Soles and palms should be checked during a general physical examination. If a pigmented or nonpigmented lesion with the diagnostic criteria suggesting ALM is discovered, it should be biopsied[17]. Saida and Yoshida recommend that any pigmented lesion on the sole of the foot greater than 7 mm in diameter should be biopsied[18]. This biopsy should be wide and deep enough for the pathologist to be able to interpret the histologic findings.

All melanomas are increasing in incidence. ALM is elusive and proportionately forms a greater percentage of melanomas in non whites. Its prognosis is worse than for other types of melanoma. Only a careful physical examination, a high index of suspicion and early diagnosis will lower its high mortality rate.

ACKNOWLEDGEMENTS

Fig 4—Courtesy of American Academy of Dermatology.

Fig 5—Courtesy of Evan Farmer, MD, Director, Department of Dermatology. Johns Hopkins School of Medicine.

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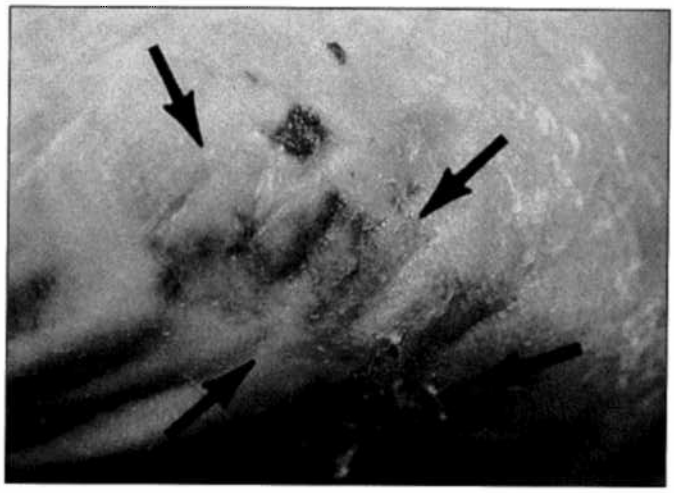

Fig. 4 - Plantar ALM, relatively early stage.

Fig. 5 - ALM of hand, dorsomedial extension.

and organized medicine to be prepared in the event of impending war at a time when Hawaii was a territory of the U.S., way out in the Pacific ocean and when communication between Washington and the Pacific military outposts of Pearl Harbor, Schofield Barracks and Wheeler Air Force Base was rather erratic and slow. This bit of medical history of that era, now 50 years behind us, has never received much mention heretofore.

It was not possible to have this published by Pearl Harbor's 50th anniversary last year, despite Rod's efforts, but it will celebrate the 51st remembrance. Rod West is to be commended for his considerable contribution to Hawaii's history and for this accolade to that generation of Hawaii's physicians.

J I Fredrick Reppun MD
Editor